LETTERS TO THE EDITOR

The moving ear syndrome: a focal dyskinesia

Although segmental dystonia of the cranial and upper limb muscles is well recognised, restricted and isolated dystonic movements of cranial musculature such as the muscles of the pinna are extremely uncommon. Dystonic movements of tranquil muscle groups such as "belly dancer's dyskinesia" (dyskinesia of the abdominal wall), an axial torsion dystonia, and four cases of "moving ears" have been reported including two patients with unilateral involuntary twitching of the ear.[1 2] We report a further two cases of unilateral movement disorder affecting the ear, one patient responding well to local injections of botulinum toxin.

Patient 1, a 23 year old white warehouseman complained of twitching of his right pinna since January 1994. Within three hours of development of the involuntary movement he experienced right temporal pain and a fluttering noise in the left ear. There was no family history of any neurological disorder. The patient had no history of any serious illnesses in the past and was not on medication.

There was a continuous semirhythmic contraction of variable amplitude at a rate of 80/min involving both ears and the scalp muscles above the ear. The involvement of the ear was more pronounced on the right. There was no palatal tremor or other dyskinesiae. Electromyography from the frontalis and auricularis superior muscles showed normal motor units firing in bursts. Brain MRI, CSF examination, and blood tests were normal except for an unexplained eosinophilia. The patient obtained considerable benefit from oral clonazepam.

The second patient, a 32 year old right handed man of West Indian extraction (born and raised in the United Kingdom) presented in April 1994 with a 12 year history of involuntary movement of the left ear. In 1983, he was thought to have a schizophrenic illness and was prescribed several forms of depot neuroleptic preparations (chlorpromazine, fluphenazine, flupenthixol decanoate) on which he has been maintained ever since.

The patient had been aware of abnormal movement of the ear at least a year before his schizophrenic illness and neuroleptic treatment. He had sought attention recently because the movements had become worse, were visible to others, and caused social embarrassment.

There was a slow rhythmic left ear elevation and retraction with occasional periods of posterior rotation and movement of the left temporalis muscle. There was no evidence of any preceding or subjective symptoms and the movements could not be suppressed voluntarily. There were no abnormal movements of the right ear or of the facial or frontalis muscles and he did not have palatal tremor or any other dyskinesiae.

Routine blood tests, serum copper studies, and lysosomal enzyme screen were negative. Electromyography of the left auricularis superior and posterior muscles disclosed synchronous bursts of normal motor unit potentials lasting 200–300 ms at a frequency of 2 Hz. Recordings from the ipsilateral frontalis muscle and the right pinna were normal. Brain MRI was normal. Injection of botulinum toxin type A (40 mouse units, Dysport, Speywood Pharmaceuticals Ltd) into the auricularis superior and posterior muscles gave appreciable benefit.

These two patients further illustrate the phenomenon of moving ears as a manifestation of focal dyskinesia. The nature of the movement disorder in these patients merits discussion.

The movements, particularly in patient 1, had a jerky element, thus raising the possibility of segmental myoclonus and a relation to palatal tremor/myoclonus. Auricular myoclonus has been described in one patient, suggesting a central origin.[3] Patient 1 had movements of the upper face involving the frontalis bilaterally and this may occur rarely in palatal myoclonus.[4] However, isolated ear movement as part of palatal myoclonus is unknown and neither of our patients had visible palatal myoclonus or heard ear clicks. Furthermore, MRI in patient 1 excluded a brainstem lesion.

In patient 2, the movements are unlikely to be a form of tardive dyskinesia as the patient was aware of the movement disorder before starting neuroleptic drugs. Ten cases of "ear wigglers" due to tics of the ear were described by Keshavan.[5] However, ear tic is unlikely in this patient as the movements were slow, rhythmic, and not suppressed by voluntary muscle contraction. In our patients, the slow often sinuous movements of the ear with a superadded jerky element are suggestive of focal dystonia with myoclonic jerks.[6 7] Also the presentation with a focal non-progressive movement disorder in adulthood is suggestive of dystonia. The reasonable responses to clonazepam in patient 1 and botulinum toxin injection in patient 2 suggest that the dystonic nature of these movements may be helped by standard treatment strategies for focal dystonia.

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1 Ilceto G, Thompson PD, Day BL, Rothwell JC, Lees AJ, Marsden CD. Diaphragmatic flutter, the moving umbilicus syndrome, and "belly dancer's" dyskinesia. *Mov Disord* 1985;5:15–22.
2 Caviness JN, Gabellini A, Kneebone CS, Thompson PD, Lees AJ, Marsden CD. Unusual focal dyskinesias. The ears, the shoulders, the back and the abdomen. *Mov Disord* 1994;5:531–38.
3 Kirk A, Heilman KM. Auricular myoclonus. *Can J Neurol Sci* 1991;18:503–4.
4 Deuschl G, Mischke G, Schenck E, Schulte-Monting J, Lucking CH. Symptomatic and essential rhythmic palatal myoclonus. *Brain* 1990;113:1645–72.
5 Keshavan MS. The ear wigglers: tic of the ear in 10 patients. *Am J Psychiatry* 1988;145: 1462–3.
6 Riley DE, Lang AE. Movement disorders. In: Bradley WG, Daroff RB, Fenichel GM, Marsden CD, eds. *Neurology in clinical practice.* Vol 2. Boston: Butterworth-Heinemann, 1991:1577–601.
7 Fahn S. Concept and classification of dystonia. *Adv Neurol* 1988;50:1–8

Acute anterior horn cell disease resembling poliomyelitis as a manifestation of respiratory syncytial virus infection

Respiratory syncytial virus (Paramyxoviridae family) is an infectious agent of remarkable interest as it is the major cause of lower respiratory tract disease in young children. It can also cause infection in adults, although it is not so severe and does not have as much epidemiological importance as in infants.[1] Despite a high prevalence of respiratory syncytial virus infection, examples of neurological disease with a causal relation have rarely been reported.[2–4] Our patient developed an acute flaccid tetraplegia preceded by a meningitic phase with serological evidence and positive cultures of a respiratory syncytial virus infection.

A previously healthy 28 year old man was admitted to hospital because of fever, meningism, and progressive weakness of the extremities. The patient had been vaccinated against poliomyelitis in 1966. A week before admission he developed an acute lower respiratory tract disease; four days later he began to have headache and diffuse weakness of all four extremities, proximal greater than distal. His 3 year old son had presented with a respiratory infection a week before the onset of the father's symptoms. Examination showed a temperature of 38·7°C, signs of meningeal irritation, and proximal weakness of the limbs (grade 4–5). Cranial nerves were intact. The tendon reflexes were hypoactive in both triceps and absent at the biceps and knees; the ankle jerks were normal. Plantar responses were flexor and no sensory abnormality was detected. His CSF had 70 white cells/mm^3 (90% lymphocytes), 1·5 g/l protein, and 66 mg/dl glucose (103 mg/dl in serum). On the second day in hospital he developed a progression of weakness with concomitant deterioration of respiration which required assisted ventilation. After 10 days sporadic fasciculations were seen in the upper extremities preceding the onset of a pronounced atrophy in all muscle groups and specially in the territory of C3 to C6 myotomes. Routine studies of blood and urine gave normal results. Tests for urinary porphobilinogen δ-aminolevulinic, and anti-GM_1 ganglioside were negative. On the ninth day in hospital CSF examination showed 200 leucocytes/mm^3 (95% lymphocytes), 3 g/l protein, and 77 mg/dl glucose. Antirespiratory syncytial virus antibody titres of 1/400 in serum and 1/1 in CSF were detected by direct immunofluorescence. Twenty five days later titres had increased to 1/1000 in serum and 1/10 in CSF. In addition the respiratory syncytial virus from CSF and bronchial aspirate samples was cultured in VERO and MRC-5 cell lines and identified using direct immunofluorescence (Monofluo Screen RSV, Sanofi). The serological tests for other viruses and bacteria commonly associated with neuromuscular diseases were negative. The patient was treated with ribavirin (200 mg orally every

Neurophysiological results

	Day 3	Day 19	Day 90	Normal
Motor:				
Right cubital nerve				
MAP (wrist/elbow) (mV)*	9·6/8·6	2/3	3·8/4·5	6-16
Distal latency (ms)	3·4	3·5	2·9	1·8-3·5
Conduction velocity (m/s)	57·2	56	58·8	49–71
F wave	34·7	NR	33·6	<31
Right posterior tibial nerve				
MAP (ankle/popiteal fossa) (mV)†	8·2/7·9	1·8/2	6·1/6·3	6-37
Distal latency (ms)	5	5·1	5	3·9-5·1
Conduction velocity (m/s)	50·3	50·2	50	45–60
F wave	58·1	49·1	47·2	<55
Sensory:				
Right sural nerve				
SAP (μV)		5	5·2	2–19
Conduction velocity (m/s)		52·9	52·1	48–62

*Muscle action potential recorded from the hypothenar eminence. †Muscle action potential recorded from the abductor hallucis. SAP = sensory action potential; NR = no response.

eight hours for six weeks). This treatment did not improve the neurological damage but resulted in sterile CSF and bronchial cultures. We also used intravenous immunoglobulin and dexamethasone before laboratory data showed features of respiratory syncytial virus infection. Spine MRI was normal. Six months later serum immunoglobulins, IgG subclasses, C3, C4, CH_{50}; B, T, and NK lymphocyte subsets, CD4/CD8 ratio, lymphocyte blastogenic responses to both B and T mitogens, NBT test, chemotaxis, serum opsonisation capability, and neutrophil and monocyte phagocytosis were normal. The patient began a slow recovery in the fifth week and artificial ventilation was discontinued on day 44. He started to walk unaided on day 70. After 10 months the patient was transferred to a rehabilitation centre. Motor examination at that time still showed grade 2–3 strength and there was considerable atrophy in the shoulder girdle muscles.

Neurophysiological studies were performed on the third, 19th, and 90th days after the onset of neuropathic symptoms (table). The nerve conduction velocities, distal motor latencies, H reflexes, and sensory nerve action potentials were always normal. The first electrophysiological examination showed slightly prolonged F wave latencies as a principal feature. Ulnar F response was absent on the next examination. The second and third studies showed a reduction in the size of the compound muscle action potentials. Electromyography showed mild to moderate denervation in the lower limbs and considerable denervation in the upper limbs, affecting mainly the C3–C6 innervated muscles. Values of somatosensory evoked potentials were in the normal range after median and posterior tibial stimulation.

Respiratory syncytial virus infection has rarely been associated with neurological abnormalities. In a few old reports respiratory syncytial virus infection could be serologically established in cases with polyradiculitis, meningitis or myelitis.[2 3] More recently it has been documented in patients with Guillain-Barré syndrome[5] and a case of Guillain-Barré syndrome preceded by a cold with serological evidence of respiratory syncytial virus infection has been described.[4] As far as we know, our case is unique in reports of respiratory syncytial virus associated neurological disorders for two reasons: the infectious agent could be cultured from CSF and the development of purely lower motor neuron damage in a pattern similar to generalised poliomyelitis. The major findings in this case were an acute generalised weakness and amyotrophy without appreciable sensory change, sphincter disturbance, or clinical involvement of the corticospinal tract, and signs of meningeal infection. The EMG features place the likely site of the lesion at the anterior horn cell. The slowing of F waves in the first examination could be related to primary demyelination in the motor root but this feature can occur in motor neuron disease.[6] The most effective chemotherapeutic management of respiratory syncytial virus infection is ribavirin.[1] Our patient was treated with this antiviral agent after the tetraparesis was complete, but ribavirin treatment was effective in sterilising the CSF.

In conclusion, respiratory syncytial virus infection should be incorporated in the differential diagnosis of the clinical syndrome of acute flaccid paralysis with meningitis. Because respiratory syncytial virus infection can be clearly diagnosed and is a treatable disorder, its recognition is important.

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1 Hall CB. Respiratory syncytial virus. In: Mandell GL, Douglas RG, Bennet JE, eds. *Principles and practice of infectious diseases*. 3rd ed. New York: Churchill Livingstone 1990:1265–78.
2 Arrowsmith WA, Smithells RW, Congdon PJ. Temporary paralysis after infection by respiratory syncytial virus. *Lancet* 1975;i:743.
3 Cappel R, Thiry L, Clinet G. Viral antibodies in the CSF after acute CNS infections. *Arch Neurol* 1975;**32**:629–31.
4 Hayashi H, Park-Matsumoto YCh, Yuki N, Itoh T, Amakawa T, Muneyuki T. A case of recurrent Guillain-Barré syndrome preceded by different infections. *J Neurol* 1993;**240**: 196–7.
5 Boucquey D, Sindic CJM, Lamy M, Delmée M, Tomasi JP, Laterre EC. Clinical and serological studies in a series of 45 patients with Guillain-Barré syndrome. *J Neurol Sci* 1991;**104**:56–63.
6 Fisher MA. AAEE minimonograph: H reflexes and F waves: physiology and clinical indications. *Muscle Nerve* 1992;**15**:1223–33.

Hereditary defect of cobalamin metabolism (homocystinuria and methylmalonic aciduria) of juvenile onset

Disorders of cobalamin (Cbl) are caused by inadequate intake, malabsorption, impairment of the extracellular transport, or inborn errors of intracellular Cbl metabolism.[1]

Two Cbl-dependent enzymes are known: the adenosylcobalamin (AdoCbl)-dependent methylmalonyl-CoA mutase (MCM; EC 5.4.99.2) and the methylcobalamin (MeCbl)-dependent methionine synthase (5-methyltetrahydrofolate homocysteine methyltransferase: EC 2.1.1.13). Nine complementation classes of defects of intracellular Cbl metabolism or of the apoenzymes have been described.[2] A combined defect of intracellular synthesis of AdoCbl and MeCbl is a rare autosomal recessive disorder reported in about 20 patients to date. Complementation analysis has disclosed two genetically distinct groups designated cblC and cblD. Most patients present in early infancy with failure to thrive, developmental retardation, and megaloblastic anaemia.[1] Manifestation later in life is rare. In one female patient the disorder manifested at the age of 14 years as subacute myelopathy and dementia.[3] After systemic OH-Cbl treatment cognition improved rapidly whereas the myelopathy responded slowly. In another patient a disorder of the CNS occurred at 21 years of age and a diagnosis of multiple sclerosis was first made.[4] After a six year relapsing and remitting disease course an isolated defect of methionine synthase (cblG) was detected.

Here a 30 year old woman (patient 1) with a 13 year disease course of relapsing and remitting myelopathy and neuropathy due to the cblC defect and her 34 year old sister (patient 2) are reported.

At the age of 12, patient 1 complained of unsteadiness of gait and urinary incontinence for a few weeks. Her motor and mental development had been normal. There was no family history of neurological disorders. She showed signs of a spinal cord disorder with pyramidal signs, and an impaired position sense. All ancillary tests, including vitamin B12 absorption and CSF studies, were normal. She had a complete remittance with prednisolone treatment. Two years later she developed similar signs, but more severe, which lasted several weeks until almost complete remission.

At the age of 19 another relapse occurred with first signs of a neuropathy with bilateral foot drop and absent ankle reflexes. Nerve conduction testing showed a reduction in amplitude and a low normal conduction velocity. A needle EMG showed chronic denervation. Treatment with high dose corticosteroids resulted in partial improvement.

In the next two years four additional relapses occurred with progressive residual deficits and mild neuropsychiatric abnormalities. Cranial and spinal MRI and CSF studies were normal. A sural nerve biopsy showed a predominantly axonal neuropathy. During the next five months there was a progressive deterioration, with inability to walk, bladder incontinence, ascending sensory loss, severe respiratory alkalosis (pH 7·51, bicarbonate 34 mmol/l, base excess 10·9) with a severe disturbance of electrolytes (hypokalaemia (2 mmol/l, normal 3·5–5·0 mmol/l), hypophosphataemia (0·55 mmol/l, normal 0·8–1·5 mmol/l)), and respiratory insufficiency. Serum Cbl was in the